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Case Report

Intracranial Pial Arteriovenous Fistula Presenting with Hemorrhage: A Case Report

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Intracranial pial arteriovenous fistula (AVF) is a rare cerebrovascular malformation, which has a single or multiple arterial connections to a single venous channel without intervening nidus, and is different from arteriovenous malformation (AVM). We report on a case of a surgically treated pial AVF. A 15-year-old girl with an altered mental state was brought to our hospital. Computed tomography (CT) showed a subcortical hematoma of approximately 24 ml in her right temporal lobe. Cerebral angiography showed an AVF supplied by the right middle cerebral artery with early drainage into the Sylvian vein and the vein of Labbe. She underwent surgical treatment with feeding artery obliteration using a clip and hematoma removal. The patient was discharged without neurologic deficits. Despite the rarity of pial AVF, for correct diagnosis and treatment, neurosurgeons should recognize this condition. Pial AVF can be managed simply by disconnection of the shunt by surgery or endovascular treatment, and a good result can be achieved.

Keywords Intracranial, Pial, Arteriovenous fistula

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INTRODUCTION

Intracranial pial arteriovenous fistula (AVF) is a rare cerebrovascular malformation, which has a single or multiple arterial connections to a single venous channel without an intervening nidus. Pial AVF was once regarded as a type of arteriovenous malformation (AVM) consisting of arterial feeders, nidus, and drainage veins.[1] However, current evidence suggests that the pathological characteristics, clinical presentation, and therapeutic options for pial AVF are different from those of AVM. Intracranial pial AVF is rare. Only 112 cases were reported from 1977 to 2009,[1] and only 1.6% of a series of 320 AVMs were AVFs.[2] In this report, we describe a case of pial AVF presenting with intracranial hemorrhage.

CASE REPORT

A 15-year-old girl with an altered mental state was transferred to our hospital. She complained of head-

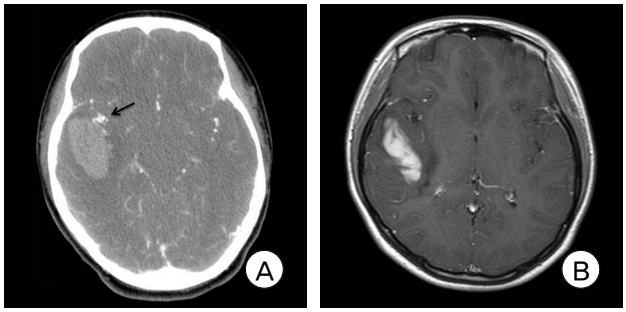

Fig. 1. Initial Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) of the patient. [A] CT angiography shows a suspicious abnormal vessel (arrow) around the hematoma on the right temporal lobe. [B] An Arterio-Venous Malformation (AVM) nidus is not observed using enhanced MRI.

ache and vomiting before her arrival, and her mental state showed an immediate decrease. Findings on the initial neurological examination indicated that she was in a stupor state (Glasgow Coma Scale was E3 V4 M5). She had no history of head trauma. Computed tomography (CT) performed for evaluation of the cause of her altered mental state showed a subcortical hematoma of approximately 24 in her right temporal lobe. CT angiogram performed in order to evaluate the cause of the hemorrhage identified a suspicious abnormal vessel around the hematoma (Fig. 1A). Magnetic resonance imaging (MRI) and cerebral angiography were performed for further evaluation of vascular malformation and to search for other causes of the hemorrhage. No nidus or mass lesion was observed in the enhanced MRI image (Fig. 1B). Cerebral angiography identified a possible AVF. The AVF was supplied by the right middle cerebral artery (MCA) and drained into the Sylvian vein and the vein of Labbe in the early venous phase (Fig. 2).

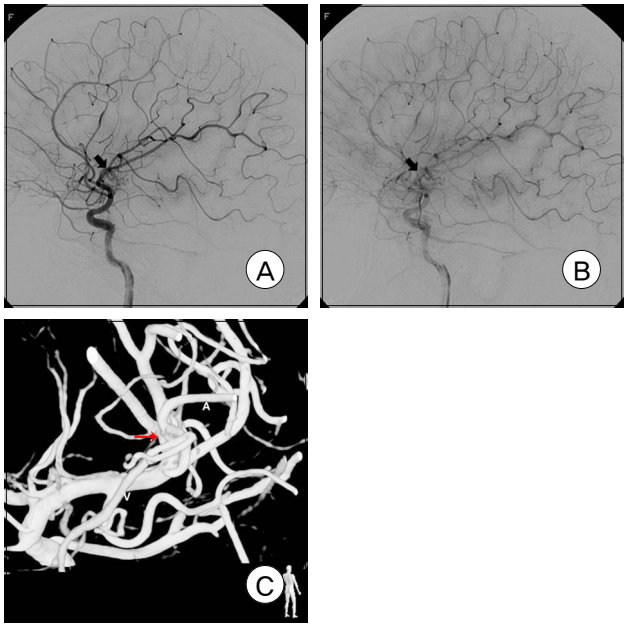

Fig. 2. Preoperative cerebral angiography of the patient. [A] A possible Arterio-Venous Fistula (AVF) (arrow) is observed around the right Middle Cerebral Artery (MCA) bifurcation. [B] The AVF drains into the Sylvian vein and the vein of Labbe in the early venous phase. [C] A 3 Dimensional (D) image shows the direct connection (arrow) of the right MCA (A) and the surrounding vein (V).

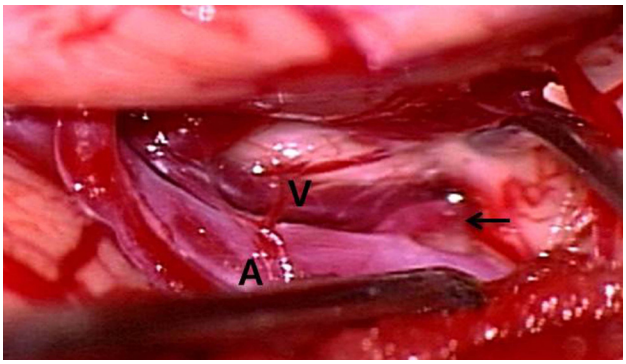

Fig. 3. Intraoperative photographs. Black arrow indicates the AVF consisting of the feeding artery (A) and drainage vein (V) show in preoperative angiography.

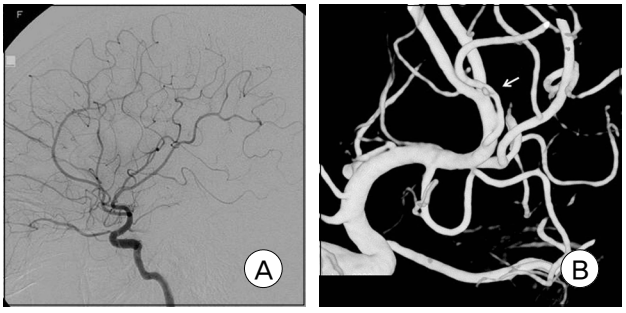

Fig. 4. Cerebral angiography one week postoperatively. [A] The early draining venous flow through the Sylvian vein and the vein of Labbe disappeared. [B] A 3D image shows that the direct connection of the artery and vein is disconnected (arrow).

A right frontotemporal craniotomy and Sylvian arachnoid dissection were performed, and an AVF consisting of the feeding artery and draining vein shown in the preoperative angiography was exposed (Fig. 3). A flow similar to arterial pulsation in the draining vein was observed on Doppler ultrasonography for assessment of vessel flow. The feeding artery was closed by application of a clip; however, arterial pulsation remained in the draining vein. The dissection and removal hematoma were extended, and other small feeding arteries were observed at the base of the hematoma. Arterial pulsation in the draining vein disappeared after obliteration of the arteries by cauterization. Cerebral angiography performed one week postoperatively showed the disappearance of the early draining venous flow through the Sylvian vein and the vein of Labbe, indicating complete ob-

literation of the fistula (Fig. 4). The patient was discharged one week later without neurologic deficit.

DISCUSSION

Intracranial pial AVFs are rare cerebrovascular lesions. Only 112 cases were reported from 1977 to 2009,[1)3)] and pial AVFs accounted for only 1.6% of a series of 320 AVMs.[2)] Previously, Yasargil et al., who reported the largest series, with eight cases, suggested that pial AVF was a fistulous type of AVM.[1)] However, a subsequent study revealed that pial AVF is a distinct entity from cerebral AVM, dural AVF, or other intracranial vascular lesions because the angio-architecture, clinical course, and therapeutic options of pial AVF differ from those of the others.[4)] Unlike AVMs, pial AVFs consist of a single or multiple arterial feeders in direct connection to a single venous drainage, and they lack a true nidus. In addition, pial AVFs are supplied by pial or cortical arteries not located within the dura mater, unlike dural AVFs, which are supplied by arteries in the dura mater. Abnormalities of AVFs arise from their high-flow nature due to the direct connection between the feeding artery and the drainage vein without an intervening tangle of vessels. Venous varix is known to coexist with pial AVF, and the associated venous varices are produced by the high and turbulent flow that results from AV shunting.[5-7)]

Pial AVFs may result from head trauma or congenital or iatrogenic causes; however, their pathophysiological mechanism is uncertain. Hoh et al. suggested that abnormal angiogenesis and associated vascular growth factors and cytokines may play a role.[8)]

Symptoms and signs of pial AVF include hemorrhage, seizure, neurological deficit, headache, and bruit. In infants, symptoms may present with cardiac failure or symptoms of increased intracranial pressure, or a palpable mass with a giant varix, skull erosion, or macrocephaly.[2)9-11)] The manifestations of pial AVFs vary according to age and the presence of a varix. Patients younger than 15 years old are more likely to have symptoms related to shunting effects and are also more likely to have varix identified by angiography: conversely hemorrhage is the major presentation in older patients,[12)] as in our case. Due to the rarity of the lesions, the natural history of pial AVFs is unknown. One study reported an association of conservative management of pial AVFs with high mortality; five (63%) of eight patients who received conservative management died due to acute or subsequent fatal bleeding.[13)] By contrast, surgical or endovascular treatment showed good results.[8)] As in our case, for appropriate treatment and to avoid unnecessarily mortality, AVFs should be considered in the differential diagnosis of young patients with intracranial hemorrhage.

Treatment of pial AVFs differs from that of AVMs. In cases of AVM, because of the high risk of recurrent bleeding, complete resection of the nidus is essential. Before Hoh et al. described the treatment of pial AVFs, removal of the varix and ligation of both arterial feeders and drainage veins was performed because it was regarded as a subtype of AVM.[14-16)] Hoh et al. reported that treatment of pial AVF by simply cutting off the shunt connection through surgery or endovascular intervention was sufficient and that removal of the entire vascular malformation was unnecessary. Disconnecting the high-flow system leads to elimination of the abnormality and its accompanying elements, such as venous varices. Surgical methods that obliterate the feeding artery by clipping or coagulation and endovascular methods can be used for the flow disconnection. Endovascular treatment is considered a simple and safe option and can approach the lesion in deep sites or critical areas. However, treatment of the AVF from a transarterial approach might be difficult in cases of multiple arterial connections or high-flow feeders.[8)] Although surgical treatment is limited for deep-seated lesions, it may be appropriate for superficially located lesions or lesions with a hematoma that require removal, as in our case. In addition, compared to endovascular treatment (86.5%), surgical treatment offers a higher obliteration rate

(96.8%).[1] Expert neurosurgical and neuroendovascular teams can carefully select the safer and more effective therapeutic methods on a case by case basis, considering the patient, lesion location, and angio-architecture.

During the operation, neurosurgeons should remember that pial AVFs consist of a single or multiple arterial feeders. As in our case, although only one feeding artery may be identified using preoperative cerebral angiography, another feeding artery that is invisible on preoperative cerebral angiography may exist. In our case, other small feeding arteries of pial AVF were detected using Doppler ultrasonography due to the arterial pulsation remaining in the vein after clipping the feeding artery. The usefulness of Doppler ultrasonography in AVM operations has been stated by many physicians.[17][18]

CONCLUSION

Pial AVFs are rare intracranial vascular malformations; however, for correct diagnosis and treatment, neurosurgeons should recognize this entity. Pial AVFs can be managed simply by surgically disconnecting the shunt or through endovascular treatment, and a good result can be achieved.

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